Idiopathic scrotal calcinosis: A rare entity and a review of the literature

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Abstract

Scrotal calcinosis is a rare condition with presentation including intradermal nodules varying in size and number. Differentials include calcification of epidermal or pilar cysts noted by the presence of keratinaceous debris. We present 2 cases of scrotal calcinosis at our institution.

Introduction

Idiopathic scrotal calcinosis (ISC) was first described in 1883 by Lewinski.[1] The clinical presentation of ISC includes gradual growth of brown-yellow, firm solitary or multiple nodules on the scrotal skin. This rare, benign condition of uncertain etiology typically begins in adolescence or early adulthood and occurs in the absence of abnormalities in calcium and phosphate metabolism.[2] The intradermal nodules tend to increase in size and number over time and can produce a white, chalky material. While these lesions are usually asymptomatic, some patients report pain and itching and there have also been reports of infection associated with ISC. Indications for surgery include relief of symptoms and preservation of scrotal cosmesis. The only treatment recommended for ISC is surgery, which allows for pathologic confirmation of the disorder.[3] We present 2 cases of this rare pathology treated surgically, as well as review of the current literature.

Case 1

A 22-year-old male was referred to the urology clinic with a 2-year history of asymptomatic scrotal nodules that had gradually increased in size and number. He denied history of scrotal trauma; he also had no history of any metabolic, systemic, neoplastic or autoimmune disease. The increasing number and size of the nodules were affecting his quality of life. He reports that he has avoided intimate relationships because he is embarrassed of his scrotal lesions. He also described increased itching secondary to the lesions. On physical examination, the patient was healthy and in no acute distress. The only significant findings on exam were multiple palpable dark yellow and brown subcutaneous nodules on the scrotum with no pain on palpation. The calcified nodules involved most of the entire scrotum (Fig. 1). His clinical findings were consistent with the working diagnosis of ISC. These lesions were confined to the scrotum with no other skin lesions elsewhere. His testes were palpably normal. Laboratory evaluation revealed a normal serum calcium, phosphorus and parathyroid hormones level.

The patient was taken to the operating room where general anesthesia was administered. The involved scrotal skin was excised using multiple elliptical incisions to preserve some uninvolved skin to allow for adequate coverage for skin closure. The scrotal skin excised did not involve the dartos fascia. Several smaller calcifications were also excised individually. All of the defects were closed using 4-0 chromic suture in an interrupted fashion. The patient has not experienced any postoperative complications to date and he has fortunately had a good scrotal cosmetic outcome (Fig. 2). Pathologic analysis revealed extensive large and small dermal calcium deposits with fibrosis, epidermal hyperplasia and focal transepidermal elimination of calcium consistent with ISC (Fig. 3). The specimens were sectioned to reveal multiple white to light yellow amorphous material within the nodular lesion.

Case 2

A 29-year-old male presented with a 15-year history of asymptomatic scrotal cysts, with no increase in size or drainage during these years. On physical examination the patient was healthy. The only significant findings on exam were multiple palpable subcutaneous nodules involving the right hemi scrotum (Fig. 4), with no pain on palpation.

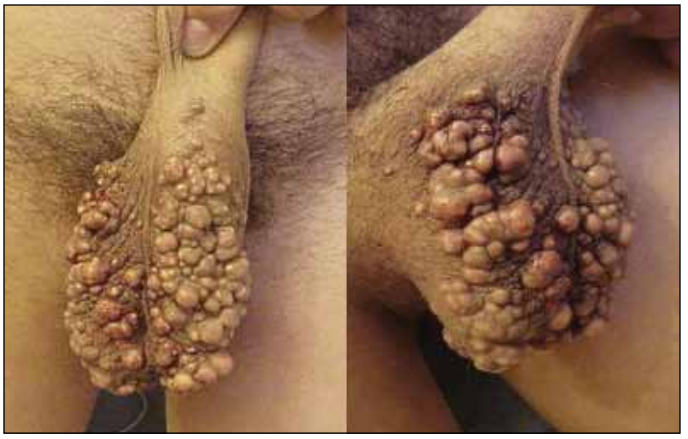

Fig. 1. Preoperative aspect: multiple subcutaneous nodules within the scrotal wall.

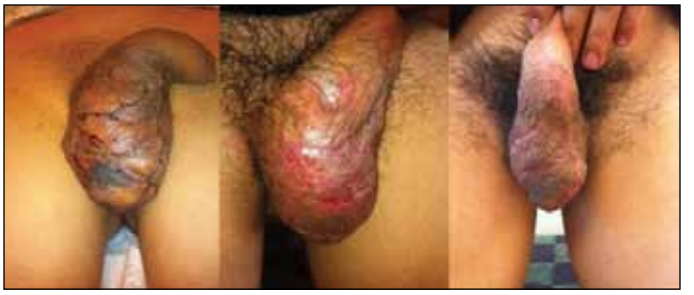

Fig. 2. (Left) surgical aspect after 3 days; (Middle) 1 month after; (Right) 3 months after the procedure.

No other lesions were discovered on the physical exam. Laboratory evaluation revealed a normal serum calcium, phosphorus and parathyroid hormones level. A surgical excision was performed under general anesthesia. The involved scrotal skin was excised using an elliptical incision above the level of the dartos muscle and the defect was closed using 4-0 chromic suture in an interrupted fashion. The patient has not experienced any postoperative complications. Pathologic analysis revealed multiple calcified nodules of variable size in the dermis, some with focal giant cell reaction consistent with scrotal skin calcinosis.

Discussion

ISC is a rare, yet benign, disease of the scrotal skin that presents with solitary or multiple, typically asymptomatic calcified nodules on the scrotum. ISC usually presents in the third decade of life, though patients commonly present years or decades after the initial onset without any systemic disorder of the calcium-phosphorus metabolism.[4] It is characterized by slow-growing yellowish-white nodules, consisting of deposits of calcium and phosphates, within the scrotal skin. The nodules vary in number, and can be solitary or grouped. These lesions are usually firm and asymptomatic, although itching or pain, episodes of infection, and chalky white exudative material have all been reported.[5] It may

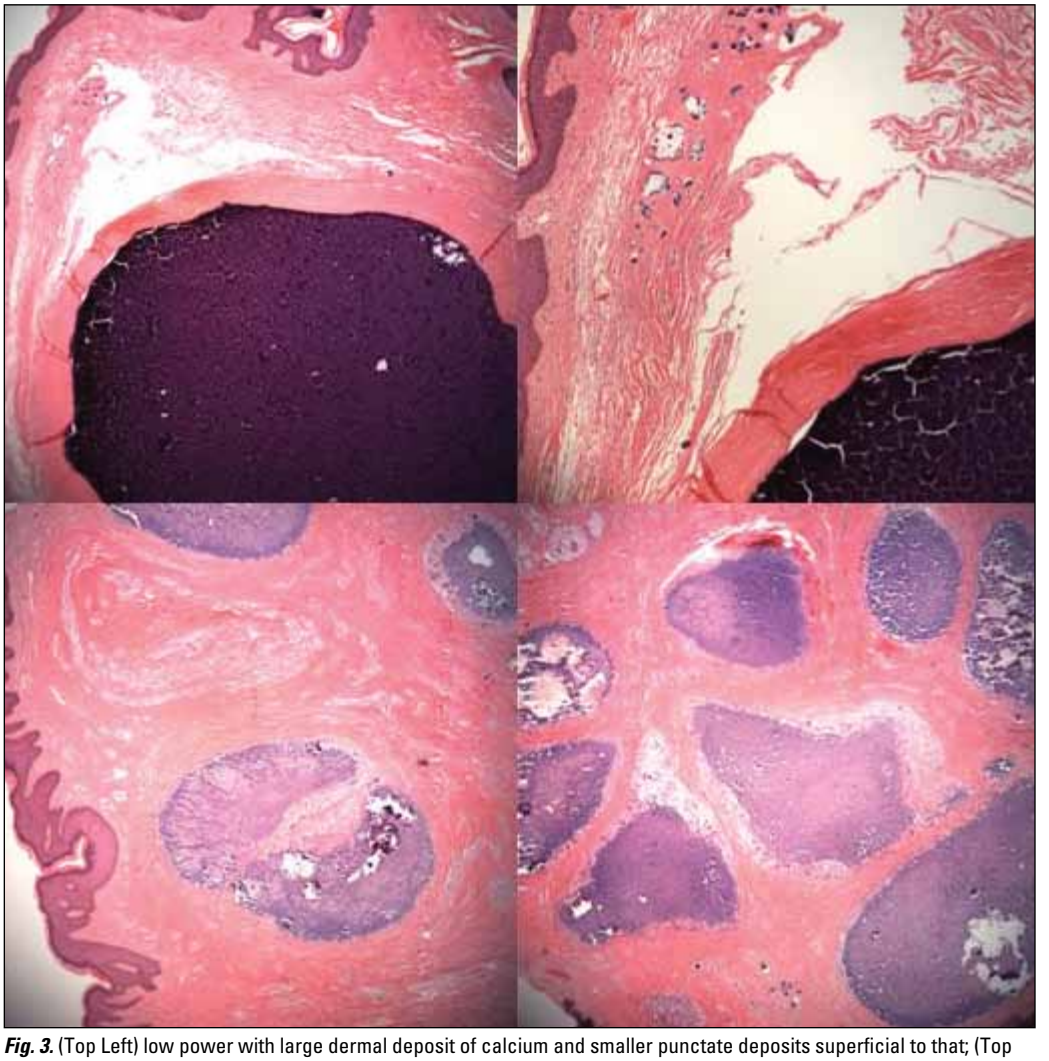

Fig. 3. (Top Left) low power with large dermal deposit of calcium and smaller punctate deposits superficial to that; (Top Right) high power to demonstrate large dermal deposit; (Bottom Left) multiple large dermal calcium deposits at low power; (Bottom Right) multiple large dermal calcium deposits at high power.

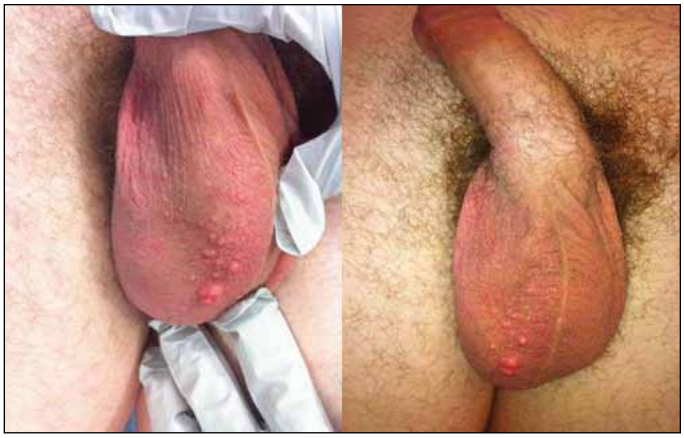

Fig. 4. Preoperative aspect: multiple cysts involving the right hemi scrotum.

considerably affect a patient's quality of life. Unusual presentations include pedunculated forms and perineal/suprapubic pain consistent with chronic prostatitis.[6] Despite its rarity, unknown etiology and benign behavior, the risk of recurrence is controversial. Some clinicians believe that all patients with ISC should undergo surgical intervention, while others disagree with surgical excision given the high probability of ISC recurrence.[7]

Controversy also exists regarding the pathogenesis of ISC. There is considerable debate as to whether this term accurately applies, as some investigators suggest that ISC is truly a late presentation of epidermal inclusion cysts that have undergone dystrophic calcification.[8] Numerous theories about the pathogenesis of ISC have been proposed. Previously, these lesions have been attributed to sebaceous cysts, calcified steatocystoma, fibroma, atheroma and xanthoma.[8,9] More recently, it has been suggested that they are the result of dartos muscle necrosis and degeneration with resulting dystrophic calcification of the dartos muscle in a process similar to the calcification of uterine leiomyoma.[10]

There is a paucity of data related to this rare disease. In 2010, Karaca and colleagues reported 2 cases discussing its possible etiology and treatment.[11] In 2011, Khallouk and colleagues reported a similar case of ICS, in which they took on the surgical approach.[3] In the same year, Grenader and colleagues reported an incidental finding of ICS in a patient with pulmonary embolism who presented to the emergency department.[12]

Histologically, ISC is characterized by calcium depositions of various sizes that are surrounded by a granulomatous reaction.[13] Despite the controversy about the origin of this entity, surgery still seems to be the treatment of choice and provides a good clinical outcome. The laxity of the scrotal skin, along with the decision to perform multiple elliptical excisions, allowed for good scrotal coverage and excellent cosmetic outcome.

Conclusion

ICS is a benign condition which can affect a patient's quality of life. The pathogenesis and basic origin remains controversial. Surgical excision appears to provide a good clinical outcome with good patient satisfaction.

Competing interests: None declared.

This paper has been peer-reviewed.

References

1. Lewinski HM. Lymphangiome der Haut mit Verkalklem Inhalt. *Arch Pathol Anat* 1883;91:371-4. http://dx.doi.org/10.1007/BF01925688
2. Saad AG, Zaatari GS. Scrotal calcinosis: is it idiopathic? *Urology* 2001;57:365. http://dx.doi.org/10.1016/S0090-4295(00)01007-4
3. Khallouk A, Yasami OE, Mellas S, et al. Idiopathic Scrotal Calcinosis: A Non-Elucidated Pathogenesis and Its Surgical Treatment. *Rev Urol* 2011;13:95-7.
4. Özgenel GY, Kahveci R, Filiz G, et al. Idiopathic scrotal calcinosis. *Ann Plast Surg* 2002;48:453-4. http://dx.doi.org/10.1097/00000637-200204000-00029
5. Turksoy O, Ozcan N, Tokgoz H. Extratesticular scrotal calcifications: their relationship with sports. *J Ultrasound Med* 2006;25:141-2.
6. Tsai YS, Tzai TS, Lin JS, et al. Scrotal calcinosis presenting with prostatitis-like symptoms. *Urology* 2002;59:138. http://dx.doi.org/10.1016/S0090-4295(01)01509-6
7. Ruiz-Genao DP, Rios-Buceta L, Herrero L, et al. Massive scrotal calcinosis. *Dermatol Surg* 2002;28:745. http://dx.doi.org/10.1046/j.1524-4725.2002.02022.x
8. Swinehart JM, Golitz LE. Scrotal calcinosis. Dystrophic calcification of epidermoid cysts. *Arch Dermatol* 1982;118:985-8. http://dx.doi.org/10.1001/archderm.1982.01650240029016
9. Baruchin AM, Baruffaldi Preis FW, Cavallinim M, et al. Idiopathic calcinosis of the scrotum: report of five cases and review of the literature. *Eur J Plast Surg* 1998;21:282-5. http://dx.doi.org/10.1007/s002380050096
10. Gi N, Gupta AK, Sachi K, et al. Idiopathic scrotal calcinosis – a pedunculated rare variant. *J Plast Reconstr Aesthet Surg* 2008;61:466-7. http://dx.doi.org/10.1016/j.bjps.2007.07.022
11. Karaca M, Taylan G, Akan M, et al. Idiopathic Scrotal Calcinosis: Surgical Treatment and Histopathologic Evaluation of Etiology. *Urology* 2010;76:1493-5. http://dx.doi.org/10.1016/j.urology.2010.02.001
12. Grenader T, Shavit L. Scrotal Calcinosis. *N Engl J Med* 2011;365:7. http://dx.doi.org/10.1056/NEJMicm1013803
13. Song DH, Lee KH, Kang WH. Idiopathic of the scrotum: histopathologic observations of fifty-one nodules. *J Am Acad Dermatol* 1988;19:1095-101. http://dx.doi.org/10.1016/S0190-9622(88)70278-9

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